INTRODUCTION

Rhabdomyosarcoma (RMS) is a highly malignant tumor and most common soft-tissue sarcoma of the head and neck in childhood [1]. The annual incidence is approximately 4.5 cases per million in children and 50% of cases are seen in the first decade of life [2]. RMS can occur in any part of the body except the bones. The common sites of occurrence of head and neck RMS are orbital, parameningeal and superficial. The locations of parameningeal RMS include nasal cavity, paranasal sinuses, nasopharynx, pterygopalatine fossa, middle ear and mastoid [3]. Although, orbital involvement is observed in 16% of cases compared to the parameningeal site in 10% of cases, yet the five year survival rate decreases from nearly 85% in orbital RMS to 50% for those with parameningeal tumours.

The clinical features are variable; basically appear to cluster around the site of the primary tumour, the age at diagnosis and histological subtypes [2]. RMS from paranasal sinuses presents with painless swelling, proptosis, sinusitis, nasal obstruction, epistaxis and cranial nerve palsies. Orbital RMS presents as a space-occupying lesion of the orbit. These features may pose as a diagnostic challenge as it may mimic other neoplastic and inflammatory masses. The histological subtypes of RMS have a prognostic significance with embryonal type being the least malignant and have a good prognosis. Sinonasal RMS is usually alveolar type, aggressive in nature with a risk for intracranial metastasis. Sinonasal RMS usually carries a poor prognosis and the risk of recurrence as these group is difficult to assess and resect.

Diagnostic modalities are computerized tomography (CT) and magnetic resonance scanning which helps to assess the spread of the disease. While evaluating the scans, particular attention is given to look for bone invasion and tumour extension into the cranial cavity and paranasal sinuses. Further bone scintigraphy is being used to detect osteoblastic metastasis. Certain studies have reported PET-CT to be superior at detecting bone and lymph node metastasis than CT and ultrasonography. The final diagnosis is based on immunohistochemical studies. The markers in RMS include antibodies against desmin (90%), muscle specific actin myoD1 (71-91%) and myoglobin. Myogenin is specific for alveolar type while vimentin and desmin are usually positive in RMS but is less specific.

RMS can occur sporadically or in association with familial syndromes with a predilection for other cancers such as Li-Fraumeni syndrome and neurofibromatosis type 1. Pediatric rhabdomyosarcoma is a highly malignant and have a good prognosis. Sinonasal RMS is usually alveolar type, aggressive in nature with a risk for intracranial metastasis. Sinonasal RMS usually carries a poor prognosis and the risk of recurrence as these group is difficult to assess and resect.

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